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RESEARCH ARTICLE

UREMIC LEONTIASIS OSSEA AN UNCOMMON MANIFESTATION IN END-STAGE RENAL DISEASE SECONDARY TO HYPERPARATHYROIDISM

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Abstract

Skeletal changes, which are commonly associated with renal osteodystrophy is a frequent outcome for individuals suffering from chronic renal failure. A rare and severe form of renal osteodystrophy known as uremic leontiasis ossea is characterized by the excess growth of the calvarial and facial bones. Evaluation with computed tomography is helpful for the recognition and treatment planning of this uncommon manifestation. We report a case of uremic leontiasis ossea in a 39 year old male who presented with swelling in front of maxilla since one month and history of tooth brush trauma with pus discharge from the upper alveolar region. The diagnosis was established by the clinical history and characteristic radiological imaging features.

Introduction:-

Leontiasis ossea is a name given to group of disorders affecting a patient's calvarial and facial bones, its other name is "lion's face" [1]. Uremic leontiasis ossea (ULO) is a serious and uncommon variant of leontiasis ossea characterized by the distinctive abnormal growth of calvarial and facial bones as a result of uncontrolled hyperparathyroidism in patients with chronic renal failure [1, 2]. Numerous complications may eventually arise from the facial cranial structures due to the progressive expansion and bone overgrowth [3].

Case Presentation

A 39-year-old male came with complaints of swelling in front of the maxilla for one month with a history of toothbrush trauma and pus discharge from the upper alveolar region. This patient was a known case of chronic renal failure (stage V) on maintenance hemodialysis. A further probe into his past revealed that he was a known case of chronic hypertension.

Ultrasound (USG) of the abdomen was taken for the patient which revealed bilateral contracted kidneys, in keeping with the end-stage renal disease.

Following a parathyroid panel, the following findings were obtained, Serum alkaline phosphatase and PTH were increased, inorganic phosphate was elevated, and serum calcium levels were on the higher side of normal (Table 1)

Parameter	Results	Reference range
Parathyroid hormone	3598 pg/ml	16-76 pg/ml

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Serum calcium	10.1 mg/dl	8.3-10.1 mg/dl
Inorganic phosphate	6.7 mg/dl	2.7-4.7 mg/d
Serum alkaline phosphatase	599 U/L	35-125 U/L
25-Hydroxy Vitamin D	15 ng/ml	10-52 ng/ml

Table 1:- Patient's laboratory results: Parathyroid hormone(PTH), serum calcium, inorganic phosphate, serum alkaline phosphatase and 25-Hydroxy Vitamin D at the time of presentation. Standard range is given.

Given the patient's current clinical history, CECT maxilla was done for the patient and it revealed loss of diploic space in the skull vault with bony expansion and loss of corticomedullary differentiation involving facial and cranial bones (Figure 1), Multiple brown tumours were also seen involving facial and cranial bones (Figure 2), Large expansile lytic lesion with hypertrophy of the anterior aspect of the maxilla in midline was causing significant separation of the teeth. Serpiginous, striated pattern of alternating high and low attenuation replacing the usual trabeculations with a lack of corticomedullary distinction noted within this expansile lesion (Figure 3).

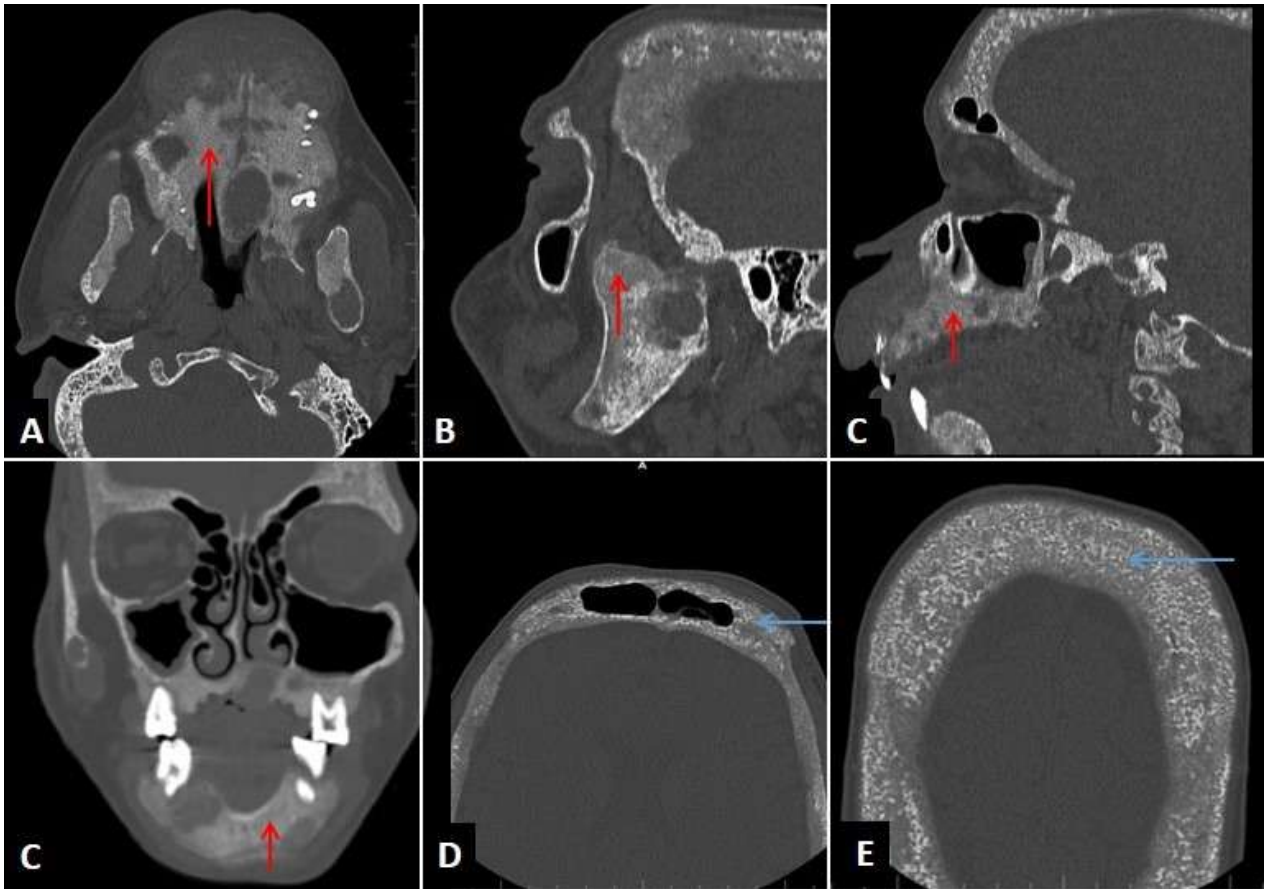

Figure 1:- Axial (A, D, E), sagittal(B) & coronal(C) computed tomography bone window. The images show bony expansion with loss of corticomedullary differentiation involving facial and cranial bones(red arrows) and loss of diploic space in the skull vault (blue arrows).

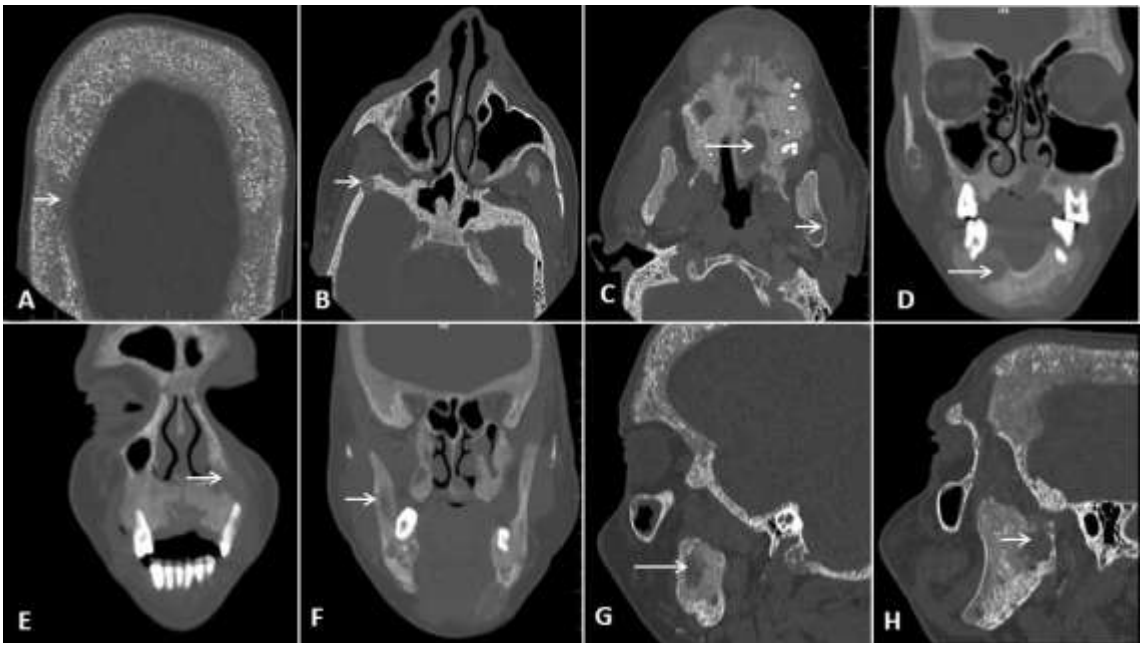

Figure 2:- Axial(A, B, C) sagittal(G, H)andcoronal(D, E, F) computed tomography bone window sections. The image shows multiple brown tumours involving facial and cranial bones (white arrows).

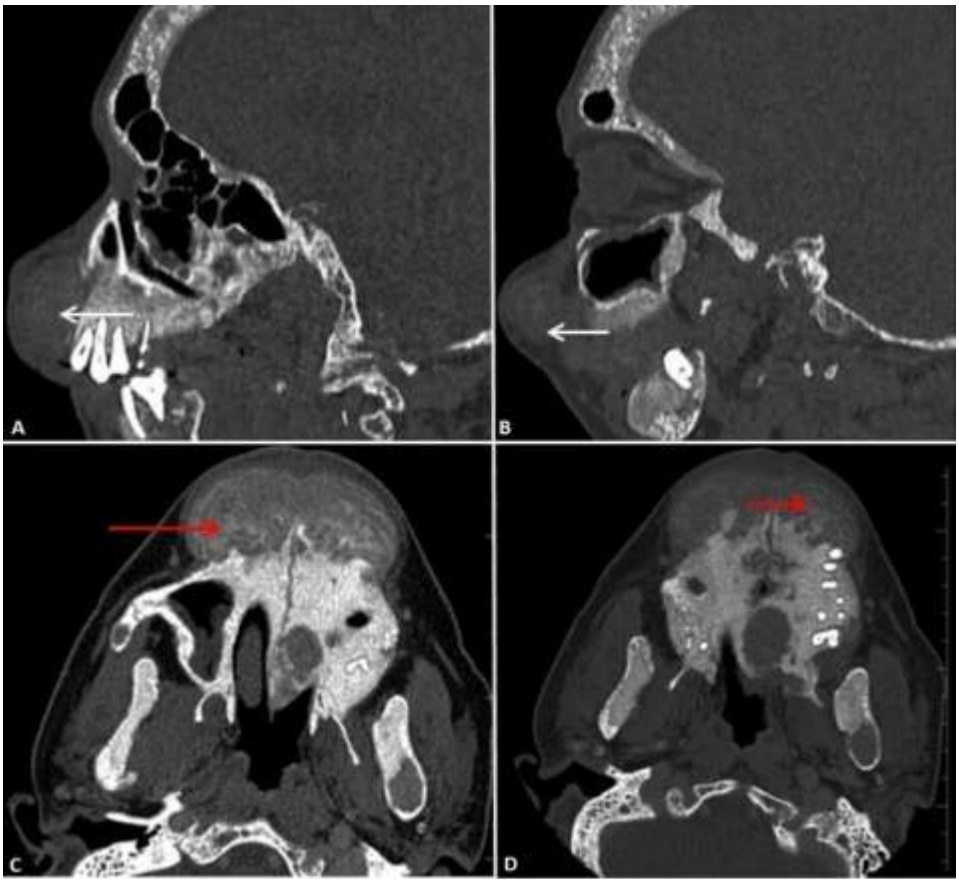

Figure 3:- Sagittal(A, B) and axial(C, D) computed tomography bone window show a large expansile lytic lesion (white arrow) with hypertrophy of the anterior aspect of the maxilla in the midline. The serpiginous, striated pattern of alternating high and low attenuation (red arrow) replacing the usual trabeculations with a lack of corticomedullary distinction is seen within this expansile lytic lesion.

Overall due to the expansion of the central aspect of the maxilla and thickening of the craniofacial bones, a leontiasis ossea-like picture is appreciated (Figure 4).

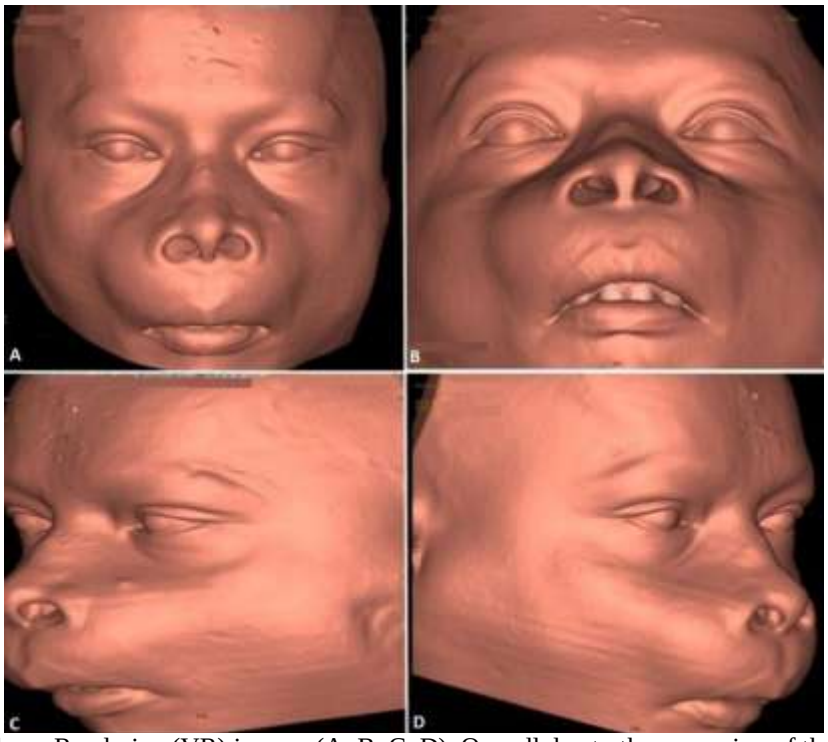

Figure 4:- 3D Volume Rendering (VR) images (A, B, C, D). Overall due to the expansion of the central aspect of the maxilla and thickening of the craniofacial bones, a leontiasis ossea-like picture is appreciated.

Skeletal survey of the patient was done which revealed, vertebral body endplates sclerosis (Figure 5), subperiosteal resorption of the middle phalanges in its radial aspect and acro-osteolysis (Figure 6) and diffuse osteopenic bones with pepper pot appearance of the skull (Figure 7).

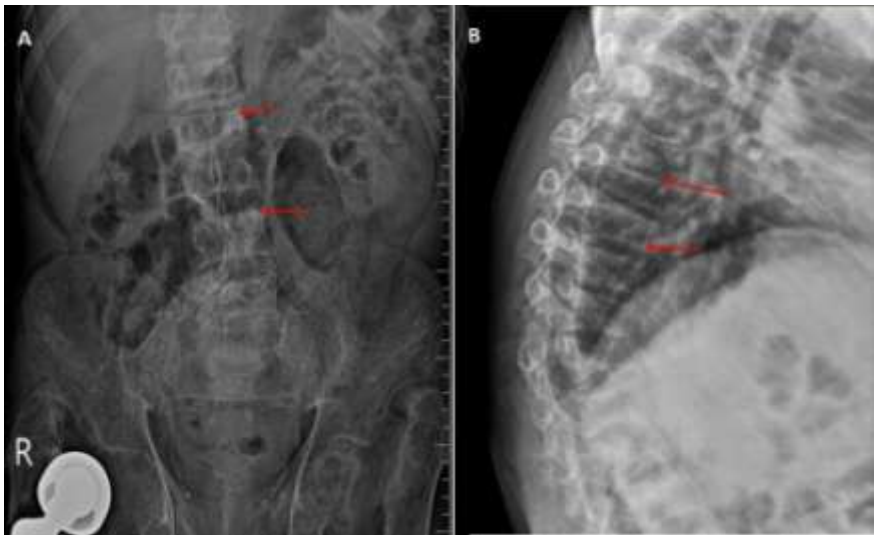

Figure 5:- X-ray frontal (A) and lateral (B) projections of the dorso-lumbar vertebra, Image shows sclerosis of the vertebral body endplates (red arrows).

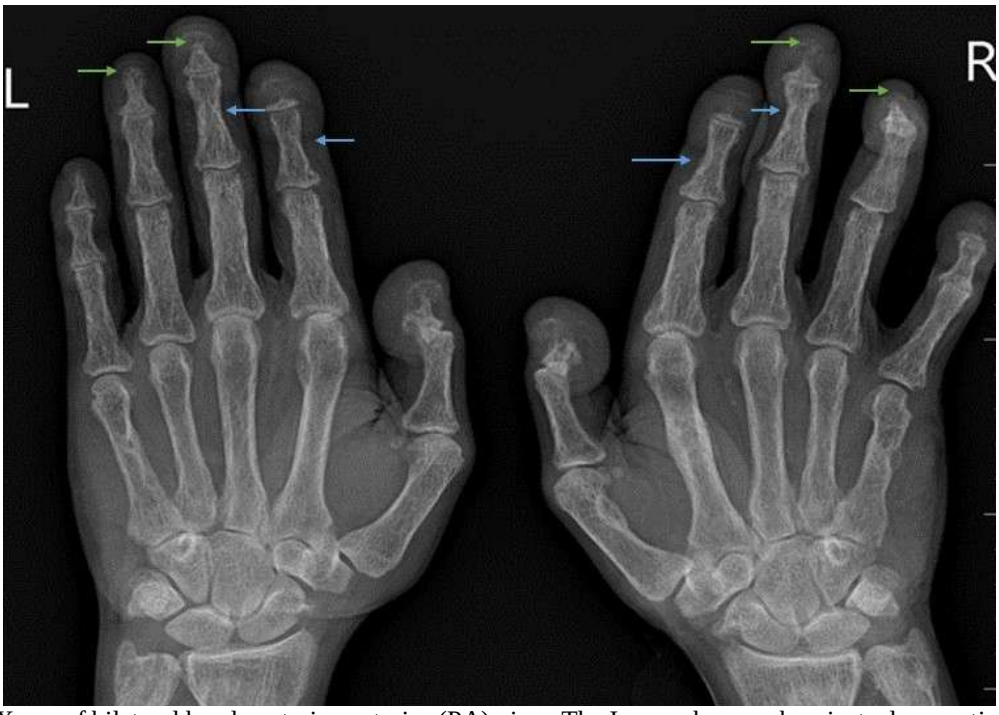

Figure 6:- X-ray of bilateral hand posterior-anterior (PA) view, The Image shows subperiosteal resorption on radial aspects of the middle phalanges(blue arrows) and acro-osteolysis(green arrows.

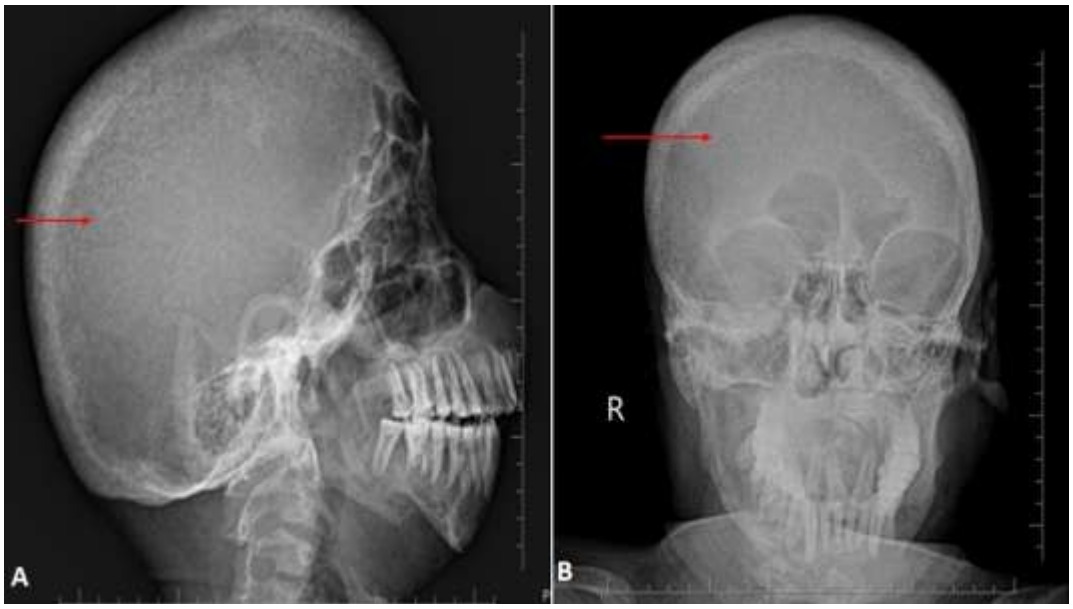

Figure 7:- X-ray skull lateral (A) and frontal (B) projections showing diffuse osteopenic bones with pepper pot appearance(red arrow).

Diagnosis of ULO was made based on the patient's clinical features, laboratory values, and unique imaging features. The patient was managed conservatively with regular hemodialysis, oral calcium and vitamin D supplements.

Discussion:-

A relatively uncommon medical condition termed leontiasis ossea (LO) is best described by excessive growth of the craniofacial bones. It is rarely reported these days, most likely as a result of improved dialysis and improved medical management of secondary hyperparathyroidism [3].

Three distinctive imaging patterns of facial bone variations are observed in patients with hyperparathyroidism and renal osteodystrophy. The first one is known as osteitis fibrosa cystica which presents with expansile multiloculated osteolytic lesions primarily affecting the pelvis, clavicle, and mandible. Histopathologically it consists of a brown mass composed of peritrabecular fibrosis, haemorrhage, macrophages containing hemosiderin, and microfractures. The second type has a ground-glass-like appearance on CT and plain radiographs, similar to fibrous dysplasia. The third type is ULO. It is differentiated by severe jaw overgrowth, poor cortical bone visibility, and serpiginous tunnelling or channelling within the bone [4].

Typical etiologies of LO include craniofacial fibrous dysplasia, Paget's disease, renal osteodystrophy, and chronic hemolytic anaemia (eg, due to thalassemia or sickle cell disease), which can be distinguished based on clinical-radiologic features [4]. The bone architecture, in this case, showed internal serpiginous tunnelling and cortical resorption, which are characteristic of uremic leontiasis ossea, consistent with this patient's risk factors [5]. Moreover, symmetric, diffuse maxillofacial osseous involvement would be atypical for fibrous dysplasia or Paget's disease, and the presence of cortical resorption would be atypical for chronic hemolytic anaemia [4,5]. Brown tumor is anincreased bone turnover pathology caused by secondary hyperparathyroidism in chronic renal failure. While mild involvement of craniofacial bones may appear as "salt-and-pepper" granular osteolysis, severe cases can be expansile with various radiographic patterns including fibrous dysplasia-like ground-glass attenuation, coarse heterogeneous sclerosis and lucency, focal brown tumors and rarely the trabecular tunnelling as seen in this case.

For hyperparathyroidism, parathyroidectomy is the preferred course of treatment [6]. Nonetheless, there are contradictory reports regarding the gradual deterioration, stabilization, and partial improvement of the uremic leontiasis ossea-related facial deformity following parathyroidectomy [7]. As an alternative, patients might gain from reconstructive surgery that makes corrections. For preoperative planning, plain computed tomography with thin sections and 3D reconstruction of the craniofacial bones is enormously beneficial [8]. Ultimately, to stop severe disfigurement from prolonged untreated secondary hyperparathyroidism, early clinical and imaging recognition of incipient uremic leontiasis ossea is crucial. An accurate diagnosis can be reached through a thorough clinical-radiological evaluation without the need for invasive tests.

Conclusions:-

ULO has unique imaging characteristics, including pronounced osseous overgrowth and typical internal serpiginous tunnelling. To prevent its severe esthetic and functional impairments, it is imperative to recognize its radiological appearance and take prompt action. For a successful course of treatment, a long-term surveillance program is necessary to look for recurrence. Alterations in biochemical values can occur before the bony changes for which preventive measures can be employed, hence biochemical screening is essential during hemodialysis

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